A CASE OF SIMPLE SEROUS CYST OF THE CEREBELLUM, WITH AUTOPSY.[1]

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The case which I herewith present for your consideration is considered worthy of being reported because it was well observed during life, and yet did not admit of a positive clinical diagnosis; because it was completed by an autopsy which revealed a localization so precise as to savor almost of a physiologic experiment; and because the completed case has its value in connection with the question of cerebellar localization and the origin of cerebellar cysts.

The patient, a married woman, 31 years of age, mother of one child, came under my observation in November, 1897. The family history presented nothing noteworthy, and the patient herself had been healthy and uncomplaining as a child, and apparently well until 3 months previous to my seeing her. She was married at the age of 24, and was for a time thereafter restless and nervous, being unable to sit still for any length of time, picking her fingers, and biting her nails. This nervousness was not of long duration and two years later when she was visited, at her home in the West, by relatives, they found her to be the same as they had always known her, quiet, cheerful, and healthy. One trait of her previous life which was related to me merits special mention in view of subsequent occurrences; this was that she was never able to ride in a railway car without becoming nauseated and very soon being attacked by retching and vomiting. Aside from such railway journeys she never vomited.

In the summer of 1895 every one thought she was irritable, not markedly so, but still more than usual; she was also nervous and fussy, allowing every-day trifles to annoy and worry her. This irritability persisted and gradually became more and more pronounced until the summer of 1897, when it culminated in repeated outbreaks of anger, and in loss of self-control which was so great as to excite comment. At this time also she began to complain of feeling tired, and of vague pains in the lower part of the back. She began to lose in weight, and her body weight fell from 120 to 115 lbs. No other symptoms were present nor did she consider herself actually ill. About November 1, 1897, the first symptoms manifested themselves, which led her to seek medical advice; she then began to complain of dizziness; at first especially in the morning, shortly after rising, she experienced sensations of dizziness, feeling as though the room were turning around, then repeatedly, usually at the breakfast table, she would support her head in her hand saying that she was dizzy, that everything was swimming. After holding her head she would feel better; there was at that time no difficulty in walking, and no further loss in weight. For a week she kept up, being treated at the office of a physician, and during this time she was repeatedly examined without the discovery of any objective symptoms. The attacks of dizziness increased in frequency and intensity, and she was obliged to take to her bed.

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During the following two or three weeks nausea and vomiting set in and were of more or less frequent occurrence, always following movements of the head and never taking place when she was at perfect rest. She complained of general weakness and appeared to be emaciating. She also showed certain psychic changes, becoming petulant, irritable and wilful, cried a great deal and appeared to be purely hysterical. About the middle of November she complained, for the first time, of headache, pain which she characterized as splitting and which was not specially localized. At this time she was examined carefully by her attending physician and by an ophthalmologist, again with negative result. It is necessary to emphasise the fact that her eyes were carefully and repeatedly examined, and fundus, muscles and visual field were always found normal.

Her gait was not then tested, but a relative who is a physician stated with certainty that there was no disorder of gait prior to her going to bed about the commencement of November.

When I examined the patient on November 27 I was unable to elicit a single objective symptom beyond that of general emaciation. The result of that examination and another one made a few days later was as follows: weight 95 lbs; temperature normal; pulse 76 to 84 per minute; thoracic and abdominal organs normal; amount of urine passed in 24 hours 56 oz.; urine contains neither albumin nor sugar. Eyes: visual field, muscles, fundus, normal; pupils equal and react promptly to light and accommodation; no nystagmus; smell, taste and hearing normal; no facial involvement; no hypoglossal involvement; a general weakness of the entire body in accord with her general emaciation, was present, but no special weakness of any extremity or muscular group was discoverable, and the lower extremities showed absolutely no motor insufficiency. There was no ataxia and all her muscles were under complete control, she being able up to a certain reduced maximum, to regulate the force, and the extent of any voluntary movement. There was no tremor. The patellar reflexes were equal and lively, and sensation was normal in all its qualities. Subjectively she was weak and complained only of becoming dizzy when she raised or moved her head suddenly; once her head in a certain position, whether she was lying, sitting or standing, the dizziness passed away.

The subsequent history of the case is brief and is one of constant anorexia, gradual and marked emaciation, some headache, considerable vomiting, occasional dizziness occurring when the head was raised from the recumbent posture; and sudden death on January 11, 1898, about 9 weeks after the onset of the first distressing symptoms.

An analysis of the nurses' daily record and of my notes shows the relative preponderance of certain symptoms; thus the temperature remained throughout between 98° and 99°, usually being 98°; the pulse varied from 76 to 120; it was 84 on the majority of dates, rose to 100 on January 3, and remained above 100 until the end.

Respirations were normal as to frequency and quality during the entire time. The average quantity of urine passed was normal; there was never any polyuria, nor was any albumin or sugar found. Sleep was interfered with only on three nights, December 3, 13, and 25. Headache was complained of on 13 days, twelve of which were prior to December 26, so that the patient was practically free from headache from that date until her death. The character of the headache is noted as severe on 5 days, on each of which it was occipital and lasted all day; upon the other occasions the ache was slight, not specially localized, and passed away in a few hours. Pain or tenderness upon percussion of the head was not present. Vomiting occurred at least once, sometime oftener, on the majority of days, yet she was entirely free from vomiting on 20 days of the time here referred to; the longest consecutive period of freedom from vomiting was 5 days, while from December 29 until her death, she vomited repeatedly every day practically retaining no food and being nourished entirely per rectum. The vomiting was always projectile in character.

Her body weight on December 31 was only 84 lbs.

Her gait was again examined on January 7, and while indicative of general weakness gave no indication of swaying, staggering, or ataxia. On this day she sat up for a half hour without dizziness.

Her psychic state was peculiar in so far as during the entire time she was markedly susceptible to suggestive influence. This susceptibility may be illustrated by the following incident: On December 18 and 19 she lay all day with her eyes closed, saying that she could not open them; all her efforts to raise the lids were manifestly futile. On the 20th she was first told positively that she could keep her eyes open for five minutes, and then commanded to do so. The command was effective, and every few hours thereafter a similar order was given, the time allotted being suggestively increased; on the 22nd the disability had passed away and thereafter no trouble of this kind was experienced. The vomiting also could be influenced suggestively to a certain extent.

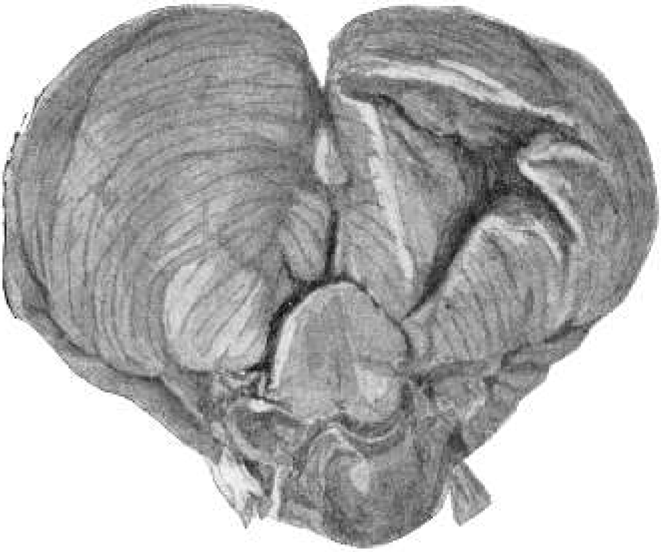

Fig. I. Cyst of the right cerebellar hemisphere.

Exitus occurred suddenly and unexpectedly; although she was very much emaciated and very weak, no indications of impending death were present.

The autopsy made 36 hours after death gave the following result: The lungs were not retracted and no adhesions were found; the left apex contained a small tubercular focus, while the right apex was perfectly normal; the heart was small, but showed nothing pathological. In the liver a number of thin walled cysts containing clear fluid were found. This was also the case in both kidneys. Each kidney contained numerous small cysts varying in size from that of a pea to a cherry; the contents were limpid in some, in others thick and syrup-like. The pelvis of the kidneys was markedly dilated the cortex of normal breadth and the papillæ shortened.

Ureters and adrenals, pancreas, intestines, ovaries and tubes normal. The stomach was dilated and empty.

The calvarium was very thick; it was easily removed and no adhesions to the dura were present. The sinus longitudinalis was empty, and the brain membranes showed no disorder. The brain itself was apparently normal in size, with smooth and unchanged vessels. The cerebellum at once gave evidence of being the seat of disease. The right half was markedly increased in size, the upper surface being prominent and fluctuating. The vermis was not deviating nor apparently compressed. An incision into the prominent fluctuating part of the right cerebellar hemisphere revealed a cyst, from which about 50 c.cm. of a clear watery fluid was evacuated. Chemical and microscopical examination of this cyst fluid revealed nothing of importance; no formed morphological elements nor hematoidin crystals were present.

The brain was then hardened for further examination. During this hardening process the whole cerebellum became badly distorted in consequence of lack of care, but the various parts and their relationship to each other were thereby not further disordered.

Upon examination after hardening the cyst was found to lie entirely in the quadrangular lobe and to have roughly the shape of an inclined pyramid, the base being in the inferior, and the apex in the superior portion of the lobe. Fig. I shows the shape and location very well. The cyst begins close to the outer margin of the flocculus and involves nearly all of the lower and internal portion of the lobus quadrangularis adjacent to the flocculus. The apex of the pyramid is in the upper portion of the quadrangular lobe opposite and external to the junction of the culmen and the declive of the monticulus. Therefore the direction of the axis of the cyst is upward, inward and backward. Thus anatomically there can be no direct communication between the cyst and the lateral recesses or the subarachnoid space.

Many portions of cyst wall were taken, imbedded in celloidin, and cut to thickness of 10 to 15 micra. The stains employed were carmine, Van Gieson and Gudden. In all more than a hundred specimens were examined.

Microscopic examinations show that at all sides the cyst is covered by cerebellar tissue. The wall is often very thin, but still the same relation prevails at all parts, all specimens showing distinct cerebellar structure external to the cyst wall.

Fig. II represents a low-power picture of the external wall of the cyst, and shows the typical relation which obtains throughout the cyst. Internally (at b) is the smooth lining cyst wall, and directly externally to it (at a) is the tissue of the cerebellum proper. This latter, of course, is slightly distorted and thinned, on account of the pressure from the distended cyst, but otherwise its structure is normal.

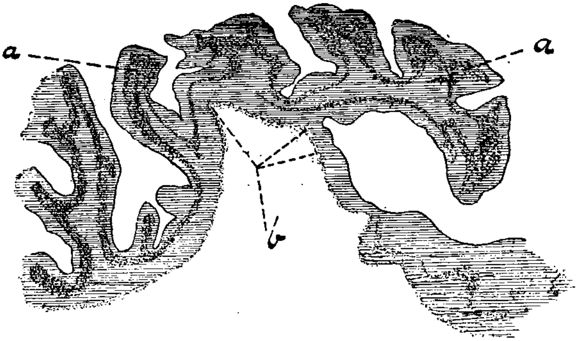

Fig. II. Topography of cerebellar cyst. *a a*, cerebellum. *b*, cyst wall.

Fig. III shows a portion of the cyst wall (viz. b of Fig. II), magnified 500 diameters. Here we have characteristic neuroglia in the cyst wall. The small cells with their comparatively large nuclei, lying in the fibrillar network are typical. At the edge of the reticulum small isolated neuroglia cells, the so-called spider cells with their processes, may be seen. These can be still further isolated by teasing the cyst wall, and such a teased specimen magnified 1,000 diameters, is represented in fig. IV.

Upon first view the cyst wall as seen in fig. III looks somewhat gliomatous, but when we note how thin the layer of cells is, and that they are not crowded together closely and irregularly, the idea of glioma formation must be given up. This is still further emphasized by the representation in fig. V. Here we have a portion of the cyst wall taken at random from the many specimens at our disposal, and here the question of tumor structure cannot arise, for it is apparent that we are dealing with simple neuroglia. We here see that all the vessels (c) are in the surrounding tissue and do not run into the cyst wall, while in gliomata we are apt to find blood vessels, at least capillaries, running into the tumor structure.

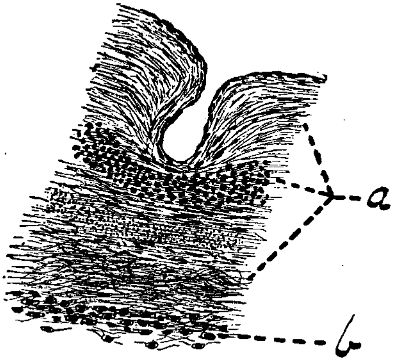

Fig. III. Portion of cyst wall magnified 500 diameters. *a*, cerebellum. *b*, cyst wall.

This is true also of all the other specimens examined, and the conclusion is thus warranted that the cyst wall is composed of neuroglia only.

The points of interest in this case appertain, firstly to the clinical picture and its relation to the pathological finding, and secondly to the nature and mode of origin of the cyst.

The clinical picture is understood when we recall that the symptoms of disease of the cerebellum may present themselves in two categories, viz., those common to any intracranial lesion, and those special to the cerebellum. In the light of modern investigations, concerning the structure of the cerebellum and its connections with other parts of the nervous system, we have learned that the cerebellum may be considered the complementary center of a reflex arc, to which the centripetal fibers from the cord ascend and from which the centrifugal fibers descend. All of these fibers pass through the inferior peduncles. Above it is connected with the optic thalamus by means of fibers which originate in the red nuclei and pass down through the superior peduncles to enter the cerebellum. These fibers are essentially efferent, conducting from the thalamus to the cerebellum, and as such are of little interest in connection with the present subject.

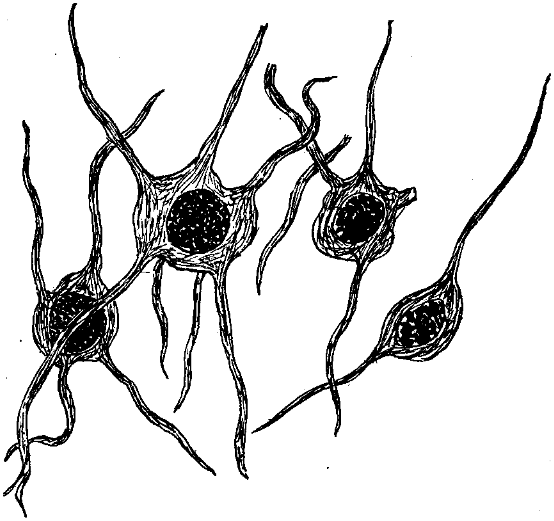

Fig. IV. Neuroglia cells teased from cyst wall.

The middle peduncles may also be here disregarded as they serve merely to connect the two lateral lobes of the cerebellum with each other.

Those tracts, however, that run to and from the cord through the inferior peduncles, and take their terminations or origin in the cerebellum, are the ones which concern us very materially.

In order to understand these different sets of fibers, it is necessary, as Bruce points out, to have a clear idea of how the fibers are given off from Deiters' nucleus. Until recently very little was known about this nucleus, whose large cells greatly resemble the large motor cells of the anterior horns of the cord, and which on account of its location above and external to the nucleus of the n. acusticus (viii) was considered as an accessory acoustic nucleus.

Although one set of fibers which originates in Deiters' nucleus does join the acoustic and terminate in the semicircular canals, these are by no means the most important fibers given off by this nucleus. Two further bundles are given off which descend to occupy certain positions in the cord; one of these sets of fibers splits up into two branches, which take their respective places in the anterior and lateral pyramidal tract, while the other set runs down, either crossed or uncrossed, to ramify in end brushes around the large motor cells of the anterior horn. It is now but a step to complete the spino-cerebellar arc; on the one hand we have the crossed fibers from the olive coursing to the dentate nucleus of the opposite side and from here to the cortex of the vermis, while upon the other hand we have two bundles of spinal fibers running to Deiters' nucleus, which in turn gives off fibers to the

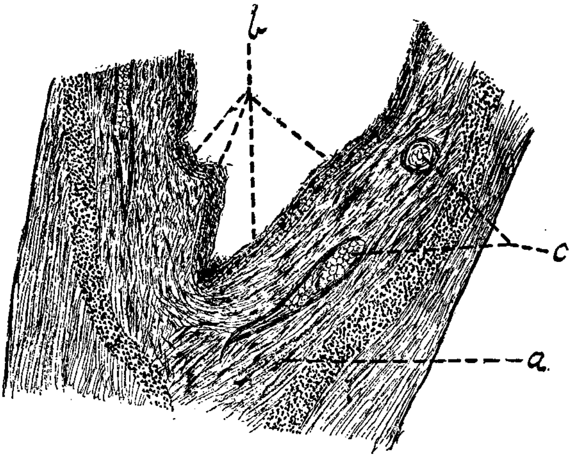

Fig. V. Portion of cyst wall. *a*, cerebellum. *b*, cyst wall. *c*, blood vessels.

roof nucleus. Interference with this spino-cerebellar tract causes no change in tactile, temperature or pain sense, but does seriously impair muscular sense.

Deiters' nucleus furthermore has two very important connections, both of which are with the motor nuclei of the ocular muscles, and of which one set of fibers has been traced to oculomotor nuclei, the other to the abducens nucleus of the same side.

We thus have centripetal impulses reaching the cortex of the vermis by way of the dentate nucleus, and centrifugal, or motor, impulses from the cortex of the vermis distributed to the ocular muscles and to the spinal motor centers for the body and extremities. From all of which it is easy to see how extensively the nucleus of Deiters is related to the various structures which surround it, and how easily any disturbance involving this nucleus could cause severe disturbances both through the spinal and ocular tracts.

Of all the structures considered, the dentate nucleus is the only one which lies in the lateral lobes; but Edinger maintains that embryologically the dentate nucleus belongs to the vermis, a fact which adds considerable significance to our preceding considerations.

The most constant and characteristic symptom of injury or disease of the cerebellum alone are: the disorder in the maintenance of the body equilibrium or so-called cerebellar ataxia; dizziness; alterations in the position of the ocular axes; nystagmic jerkings, and finally, a weakness of the musculature of the body, which in a unilateral lesion is probably present upon the side of the lesion.

In short the symptoms may be summarized as ataxia and dizziness, both of which symptoms we would, from our anatomical knowledge, expect to be present if Deiters' nucleus were involved. Interference with the ocular and spinal fibers would give us disturbances in co-ordination and consequent ataxia, while involvement of the vestibulary fibers of the eighth would cause dizziness. Now considering the location of Deiters' nucleus and its intimate relation with the surrounding structures, we may postulate that cerebellar symptoms would follow any involvement of that portion of the cerebellum lying between the two dentate nuclei, or in other words, of the vermis cerebelli. It has already been noticed, and often recorded, that very extensive lesions of the lateral lobes of the cerebellum have occurred with few or no cerebellar symptoms, and Nothnagel has pointed out that such lesions produce symptoms only when they also involve the middle lobe or vermis. Of course a tumor growth or any process causing enlargement of the hemispheres would give rise to some pressure upon the middle lobe and thereby produce some cerebellar symptoms. But Nothnagel has gone further and maintains that in any lesion causing loss of substance of the lateral lobes only, no cerebellar symptoms whatever will be found.

In our case then, the symptoms are in thorough accord with what the preceding considerations would lead us to expect. The cyst involves a large part of one lateral lobe but is confined entirely to the hemisphere, leaving the structure of the vermis free, even from pressure. All pressure which here existed must have been downward and inward so that none of the nuclei could have been implicated.

Yet dizziness, a cerebellar symptom, was present. In this connection it is necessary merely to recall the course of the eighth nerve before it makes its exit from between the transverse fibers of the pons and the external margin of the olive. Here the anterior root has already received the vestibulary fibers from Deiters' nucleus, so that pressure at any point in this vicinity would give us the vestibulary symptom. The dizziness accompanying disease or involvement of the eighth nerve causes the patient to feel as though he were being whirled through space—a sensation which may be constant or occur only in attacks. This is practically the same characterisation as that of cerebellar dizziness, and there can hardly be a doubt that the symptom is produced by a disturbance in Deiters' nucleus by way of the vestibulary tract to the acoustic nerve.

A brief retrospect thus shows us that all of the symptoms in our case fit the pathologic finding. The only symptoms present, with the exception of the dizziness, are those which would be caused by any intracranial growth or condition causing intracranial pressure; the dizziness on the other hand could have been caused by any disorder involving the middle ear, or acoustic fibers at any point in their course from the semicircular canals to their origin, and so does not necessarily indicate cerebellar disease. From all this it seems to us that no localization of the process could have been made during life. Having reconciled the clinical picture with the pathologic finding let us turn to the question of the origin of the cyst.

Various kinds of cysts are met with in the cerebellum; there are the hydatid cysts, those due to cysticercus cellulosæ, the cysts which are the result of hemorrhage or softening, and those due to cystic degeneration of neoplasms such as gliomata or sarcomata; or there may occur cystoid dilatations, eversions of the ventricles, or the transformation into a cyst, of pre-existing physiologic cavities, such as the lateral recesses, through agglutination of their walls.

Such separation from the original cavity may take place during fetal life, in consequence of developmental anomalies, while the transformation into a cyst is set up at a much later period. In addition to the foregoing there exists a comparatively rare variety of cerebellar cyst, of which a number of cases have been described and which upon post-mortem examination have revealed a cyst containing clear fluid, but giving no evidence of having been produced by any of the ways already mentioned. Such cases can be described only as simple serous cysts of obscure origin.

Our case giving no evidence of hydatids, cysticercus cellulosæ, hemorrhage, hematoidin crystals, nor tumor growth, and the absence of epithelial or endothelial cells precluding the formation from a pre-existing ventricular cavity, I can range it in no other class than that of simple serous cyst. I do this in full knowledge of the cases described by

Williamson. This observer has met with two cases in which careful dissection and fairly complete microscopic examination failed to give any evidence of new growth; yet a more extended examination revealed a very small patch of tumor formation in a part of the cyst wall. In case I of Williamson in the entire wall of the cyst, which was nearly the size of a pigeon's egg, no evidence of new growth was detected, with the exception of one spot, which proved to be a minute oval glioma measuring only 2.5 by 4 millimeters; nearly one-half of this neoplasm consisted of blood vessels.

In connection with this case Williamson himself says: "If a tumor growth can undergo cystic degeneration to such an extent that the only remaining new growth shall be of the dimensions just mentioned, it does not seem improbable that in some cases the whole of the growth may disappear and only a cyst remain."

In my case the entire cyst wall was carefully examined with a magnifying glass and nothing but a perfectly smooth cyst wall was found. Sections for microscopic examination were then taken from various parts of the cyst wall and, considering the number of sections made, practically the entire area was examined.

The fact that this examination failed to reveal any evidences of new growth does of course not answer Williamson's hypothesis of a possible complete absorption of such a growth. If, however, we add the other facts in the case, viz., the absence of all symptoms in the history pointing to the growth of a tumor, the inability to give any other satisfactory explanation as to the origin of the cyst, and the fact that the autopsy showed cysts in other organs, in the liver and kidneys, a fact which warrants the assumption of a common origin of all the cysts, then I can arrive at no more satisfactory conclusion than the one that we are dealing in this case with a simple serous cyst. Whether such a cyst may arise in consequence of dilatation of a lymph space, or whether such cystic formation is of congenital origin, as cystic kidney and liver are by some authors assumed to be, are questions which merit consideration, but cannot be decided.